

Management of a renal calculus larger than 4 cm in a patient with tuberous sclerosis complex-associated angiomyolipoma

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ABSTRACT

Renal calculi in patients with renal angiomyolipomas are difficult to treat because of the hemorrhagic potential of these tumors. We describe the case of a 65-year-old man having tuberous sclerosis complex-associated multifocal renal angiomyolipomas with a large renal calculus. The patient presented with left flank dullness and a previous history of spontaneous angiomyolipoma rupture. Intravenous pyelography revealed a 43 × 16 mm calculus in the pelvis and lower calyx of the left kidney. The calculus was successfully removed by retrograde flexible ureterorenoscopy and holmium-YAG lithotripsy. Flexible ureterorenoscopy is an effective, minimally invasive therapy for patients at high risk of renal hemorrhage.

Key words: Angiomyolipoma, flexible ureterorenoscopy, nephrolithiasis, tuberous sclerosis complex

INTRODUCTION

Tuberous sclerosis complex (TSC) is a genetic disorder that can affect almost all organ systems. Renal manifestations occur with high frequency, the most prevalent being angiomyolipoma (AML). TSC-associated AMLs tend to be larger, more numerous, and more likely to cause spontaneous hemorrhage compared with sporadic AMLs. Removal of renal calculi from kidneys with AMLs is particularly difficult because these tumors increase the potential for hemorrhage. We report the case of a 65-year-old man with TSC-associated AML and a renal calculus greater than 4 cm in his left kidney. The calculus was successfully removed by retrograde flexible ureterorenoscopy (F-URS) with holmium laser lithotripsy.

CASE REPORT

A 65-year-old man was admitted to our department with left flank dullness. He was previously diagnosed with TSC after being identified with the typical features, such as dermatological lesions (facial angiofibroma, ungual fibromas, and hypomelanotic macules on the toes), subependymal nodules, and multiple AMLs in both kidneys. While there was no family history of TSC, the patient had childhood epilepsy. He also experienced two episodes of hypovolemic shock concomitant with extensive retroperitoneal hemorrhage caused by spontaneous AML rupture, once in each kidney. Following the second episode, which occurred in the left kidney, he developed sepsis caused by *Staphylococcus aureus*, which was isolated from both venous blood and urine. He received conservative therapy without blood transfusion for both episodes of rupture.

Abdominal palpation revealed bilateral kidney enlargement without tenderness. Blood counts were within normal limits except for a low red blood cell count ($331 \times 10^4/\text{mm}^3$; normal, 430×10^4 – $570 \times 10^4/\text{mm}^3$), low hemoglobin (9.5 g/dl; normal, 13.5–17.0 g/dl), and a low hematocrit value (30.4%; normal, 40%–50%). Serum creatinine level was 1.0 mg/dl (normal, 0.6–1.1 mg/dl). Urinalysis demonstrated sterile pyuria with a negative urine culture. Kidney–ureter–bladder (KUB) radiography revealed a 43 × 16 mm renal calculus in the left kidney [Figure 1a]. Intravenous pyelography revealed that the calculus filled the pelvis and lower calyx, causing dilatation of the upper calyx [Figure 1b]. Abdominal enhanced computed tomography (CT) demonstrated a significant bilateral renal

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AML burden in addition to the renal calculus in the left kidney [Figure 2]. We decided to remove the renal calculus for several reasons, including the present clinical symptoms and a previous history of urinary tract infection. Minimally invasive retrograde F-URS with holmium laser lithotripsy was chosen because multiple renal AMLs have hemorrhagic potential.

A 6.9-Fr F-URS, a 10-Fr ureteral access sheath, and a 200- μ holmium: YAG laser fibers were used for treatment. We attempted to vaporize the stone by “painting” the laser fiber over the stone surface. A 1.9-Fr nitinol basket, which minimizes the loss of deflection during F-URS, was used to extract large fragments and reposition them from the lower pole calyx into a position more readily accessible by the laser fiber, such as the renal pelvis or upper pole calyx. Complete stone fragmentation was achieved and a 4.8-Fr double pigtail stent was inserted. Total surgical duration was 195 minutes. The stone was composed of calcium phosphate. No evidence of greater than 2-mm residual stones was observed before removal of the ureteral stent in postoperative KUB radiography. On postoperative day 5, the stent was retrieved using the tether suture. Postoperative course was uneventful with gradual disappearance of clinical symptoms. Three months after surgery, KUB radiography confirmed the absence of any residual stones [Figure 3], and urinalysis was also revealed normal findings.

DISCUSSION

Renal manifestations occur frequently in TSC. Rakowski *et al.* reported renal lesions in 57.5% TSC patients; of these, AML was present in 85.4%, cysts in 44.8%, and renal cell carcinoma in 4.2%.^[1] TSC-associated AML is more likely to cause spontaneous hemorrhage compared with its sporadic counterpart, with a hemorrhagic risk of 25% to 50%.

TSC patients are also predisposed to nephrolithiasis because of both the disease and its treatments. For example, drugs like topiramate have proven to be effective for some forms of TSC-associated epilepsy; however, they also increase the risk of nephrolithiasis. Patients are also at higher risk for renal cystic disease because disruption of distal tubular function by a significant cyst burden leads to hypocitraturia. Our patient was not undergoing any treatment for epilepsy, and CT demonstrated no evidence of renal cystic lesions.

Traditional surgical treatment of renal calculi, including minimally invasive procedures like percutaneous nephrolithotomy (PNL) or extracorporeal shockwave lithotripsy (ESWL), may lead to bleeding in patients with renal AMLs. PNL remains a milestone technique with a high success rate and with a low percentage of major complications; however, the blood transfusion rate reported varies from 11.2% to 17.5%.^[2] PNL involves puncture and dilation of the renal collecting system via the renal



Figure 1: (a) KUB radiography reveals a lower pole renal calculus in the left kidney. (b) Intravenous pyelography shows the calculus causing dilatation of the upper calyx



Figure 2: Abdominal enhanced computed tomography demonstrates significant bilateral renal AML burden and a large renal calculus in the left kidney



Figure 3: Three months after flexible ureterorenoscopy, follow-up KUB radiography confirms the absence of any residual stones

parenchyma and poses an unacceptable risk of hemorrhage in kidneys with AML, although there are two reported cases of PNL performed directly through a renal AML.^[3,4] On the

other hand, ESWL requires large numbers of shock waves, which can lead to capillary damage with parenchymal or subcapsular hemorrhage; ESWL-induced retroperitoneal bleeding has been reported in a patient with renal AML.^[5]

Recent studies have evaluated F-URS as an alternative treatment for patients with contraindications to PNL or ESWL, such as pregnant woman, obese patients, or those with coagulopathy. Indeed, F-URS using pulsed laser energy can fragment stones with less risk of hemorrhage. Furthermore, F-URS is a minimally invasive procedure that can treat <2-cm renal calculi with a low morbidity and high success rate. Our case and other reported cases indicate that F-URS can also safely treat larger stones.^[6] However, the management of lower pole renal calculi with F-URS remains debatable because of limited access and poor clearance of stone fragments.^[7] By the current literature, PNL should be considered the first line of treatment for greater than 2-cm lower pole renal calculi.^[8] Further advances in endoscopic technology and techniques are required to resolve the limitations of F-URS, including the technical difficulty in removing large stones, intrarenal influx with infected stones, and prolonged surgical duration.

In conclusion, retrograde F-URS with holmium laser lithotripsy is an effective, minimally invasive therapy for patients at high risk of renal hemorrhage.

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